Segmental Arterial Mediolysis of the Abdominal Vessel: A Case Report

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Segmental arterial mediolysis (SAM) is a rare but serious arteriopathy of unknown etiology with life-threatening manifestation. It is characterized by lysis of the medial layer of the arterial wall and may result in dissection, stenosis, occlusion, and aneurysm formation. We would like to report a case of SAM that occurred in the hepatic and visceral arteries and caused the ischemia.

Keywords: Segmental arterial mediolysis; Artery; Bleeding; Occlusion; Angiography

Introduction

Segmental arterial mediolysis (SAM) is a rare but serious non-atherosclerotic, non-inflammatory arteriopathy of unknown etiology with life-threatening manifestation (1). It mainly affects the medium-sized vessels of the abdomen and is characterized by lysis of the medial layer of the arterial wall. It may result in dissection, stenosis, occlusion, and aneurysm formation (2, 3). Given its rarity and angiographic similarities to other vasculopathy, the diagnosis of SAM can be challenging and is often missed. We here describe the case of SMA focused on CT angiography features.

Case Report

A 34-year-old female patient was referred to our hospital for recurrent abdominal pain. The pain is usually aggravated in the post-prandial period. Routine laboratory tests were within normal range, including high sensitivity C-reactive protein (hs-CRP) of 0.46 mg/dL.

The CT angiography revealed the concentric luminal narrowing of the celiac trunk and proximal superior mesenteric artery (SMA). There was no significant stenosis in both renal arteries (Fig. 1). Digital subtraction angiography for mesenteric arteries presented multifocal strictures of distal SMA, left colic artery, and sigmoid artery.

One-day after, the abdominal pain was aggravated. The second CT angiography revealed the segmental occlusion of the right hepatic artery with the ischemic change of the subcapsular area of the right posterior section of the liver. In addition, diffuse pneumatosis intestinalis was noted with markedly decreased bowel enhancement (Fig. 3).

Immunological laboratory tests were performed to exclude autoimmune or inflammatory causes of...
Fig. 1. (A, B) Dynamic CT revealed the segmental narrowing of the proximal superior mesenteric artery (SMA) and (C) multifocal strictures in the celiac trunk and proximal common hepatic artery (arrows). (D, E) CT angiography also showed multifocal luminal narrowing of the celiac trunk and SMA (arrows). An aberrant right hepatic artery from SMA was faintly delineated (black arrows). (F) Both renal arteries were intact.
vasculopathy, and the results were all within the normal range. The SAM was diagnosed based on the patient's history, clinical manifestation, angiography findings, and ruling out other causes.

Fig. 2. (A) Digital subtraction angiography for superior mesenteric artery presented strictures in the distal portion (arrow). (B, C) On angiography for inferior mesenteric artery also presented multifocal strictures of the left colic artery and sigmoid artery.
Discussion

SAM is a rare arteriopathy with life-threatening manifestations. This lesion is postulated to develop due to cytoplasmic vacuolar degeneration of arteriolar smooth muscle cells. Integration of these vacuoles then leads to disruption of the media, intramural hemorrhage, fibrin deposition at the adventio-medial junction, and granulation tissue formation (4). The most commonly affected vessels are the visceral arteries, followed by the renal, intracranial, abdominal muscular, internal carotid, iliac, pulmonary, and vertebral arteries, and multivessel involvement is common.

The hallmark of angiographic diagnosis is stenosis, occlusion, aneurysmal formation, arterial dilatation, and dissection of medium-sized vessels. Numerous lesions are often identified, affecting arteries in a segmental skip pattern, with the diseased segments demonstrating the involvement of the circumference or only a portion of the arterial wall (5). Arterial dissections account for most of the reported cases of SAM, followed by pseudoaneurysms. When dissection of peripheral arteries unrelated to the aorta is observed, a diagnosis of SAM should be considered (6).

The SAM can mimic other vascular diseases, and the discrimination of SAM is essential because the management options are vastly different. The primary differential diagnoses are atherosclerosis, inflammatory vasculitis, and fibromuscular dysplasia. Atherosclerosis typically occurs at the branch points of vessels and

Fig. 3. (A, B) Dynamic CT one day after revealed the segmental occlusion of the right hepatic artery (arrows) with the ischemic change of the subcapsular area of the right posterior section of the liver (arrows). (C) In addition, diffuse pneumatosis intestinalis was presented with markedly decreased bowel enhancement.
multiple lesions throughout the vascular tree in person with cardiovascular risk factors. SAM is usually confined to vessels of one anatomy site (7). The clinical manifestation, angiographic features, and normal laboratory results of inflammatory and autoimmune factors are helpful to differentiate inflammatory vasculitis such as polyarteritis nodosa and Takayasu’s arteritis. Another differential diagnosis with similar angiographic findings to SAM is fibromuscular dysplasia. It also presents a beading appearance. However, it is usually asymptomatic and affects younger patients with predominant involvement of the renal arteries (8). Also, in our case, both renal arteries were relatively intact.

The mortality rate of SAM is as high as 50% in patients who present with life-threatening bleeding, and early diagnosis and detection of disease progression have reduced the mortality rate to 25%. No formal guidelines for the management of SAM exist yet. Recently, catheter-based endovascular techniques can be a successful, minimally invasive treatment option in managing this potentially life-threatening condition and may also provide a temporary bailout measure in the acute phase before definitive surgical treatment at a later stage (2).

In conclusion, SAM is a diagnosis that should not be neglected in patients presenting with sudden-onset abdominal pain and abnormal angiographic findings. Although the mainstay of treatment has generally been surgical intervention, the less invasive endovascular treatment demonstrates an emerging role and is proving successful. In patients placed under disease surveillance, CTA scans at regular intervals are warranted to detect early disease progression.

References